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FAMILIAL SICK SINUS SYNDROME IN TWO SIBLINGS*

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Two siblings with sinus node disease are presented. The patients were severely affected and required permanent pacing. A cardiac electrophysiologic study was conducted in Case 2, which revealed an atrioventricular conduction disturbance in addition to sinus node dysfunction. The parents and other siblings showed no evidence of sinus node disorder. The occurrence of this disease in these siblings suggests an autosomal dominant mode of inheritance with variable penetrance. *Key words: sick sinus syndrome, familial.*

A disturbance in sinus node function is a common cause of symptomatic arrhythmias and may result from failure of sinus automaticity, or the inability to propagate impulses from the sinus node to the atrium, or a combination of both[1-4]. This type of arrhythmia rarely occurs in childhood, but is frequently associated with extensive atrial operations[5]. There are many other etiological or associated conditions. The cause of sinus node dysfunction may be classified as intrinsic or extrinsic[6-11].

In view of the multifactorial etiology of sinus node dysfunction, it is not surprising that no single underlying histological abnormality has been identified, but rather a variety of findings has been reported[1-4, 12, 13]. Among these factors, familial occurrence is rarely involved[6-11]. In this report two siblings with sinus node dysfunction are presented.

Case Reports

Case 1

A three-year-old girl was apparently well until the age of two, when she developed syncopal attacks. Physical examination revealed bradycardia and a systolic murmur which was believed to be associated with ventricular septal defect. The

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following year, she had frequent syncopal episodes and a routine examination was unremarkable except for a heart rate of 80/min, with frequent irregularity. The ECG showed basically a junctional rhythm (Fig. 1). The telecardiogram revealed slight cardiomegaly. The M-mode echocardiogram was normal. She was hospitalized at the age of three because of syncopal attacks. Exercise and the administration of atropine and orciprenaline did not significantly increase the heart rate, which confirmed the diagnosis of sinus node dysfunction. It was then decided that a permanent pacemaker be inserted in the patient. An epicardial demand pacemaker was used and continuous pacing was achieved at a rate of 100/min. Following insertion of the demand pacemaker she had no syncopal episodes. The last telecardiogram taken was normal and her last ECG showed paced rhythm with sinus node dysfunction.

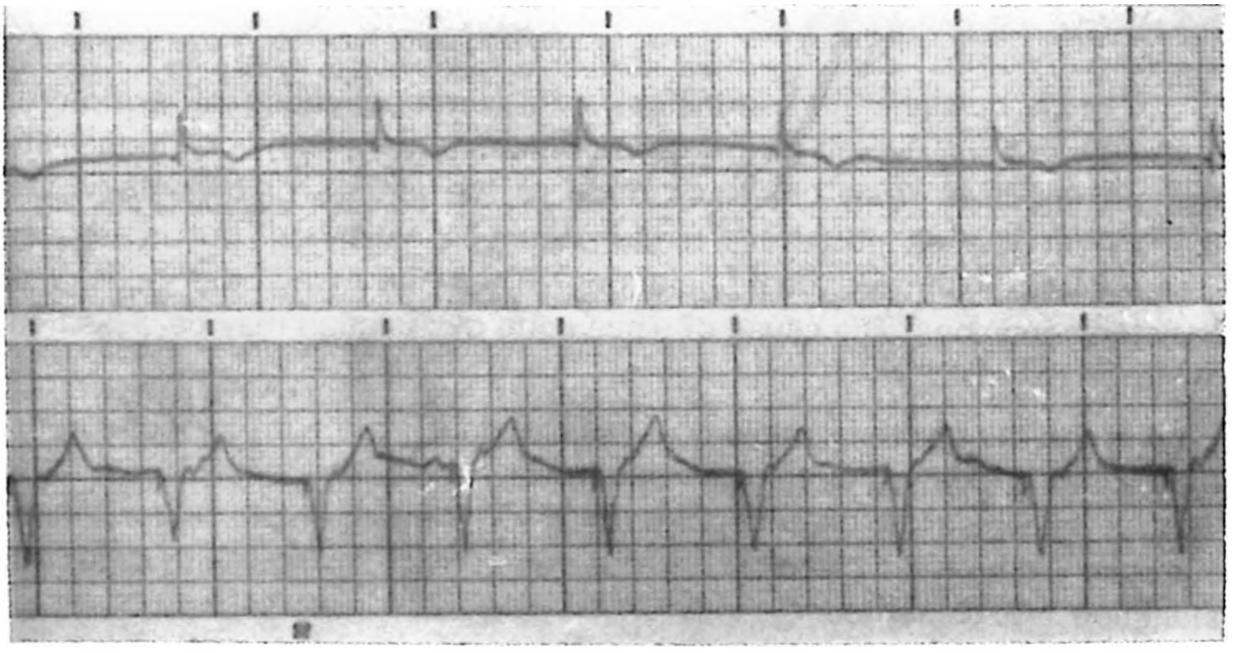

Fig. 1: Top panel shows junctional rhythm and absent P waves. The bottom panel demonstrates pacemaker rhythm.

Case 2

An 11-year-old boy was admitted to the hospital with dizziness. He had been apparently well until five months prior to admission. A routine examination revealed bradycardia, and a systolic murmur was heard in the fourth left intercostal space. The ECG revealed sinus bradycardia with frequent sinus arrest and an infranodal rhythm at a rate of 42/min (Fig. 2). On some strips a normal sinus rhythm was observed. Exercise did not noticeably increase the heart rate. The two-dimensional echocardiogram revealed left ventricular dysfunction with a normal cardiac anatomy. The telecardiogram showed an enlarged heart. Before the permanent pacemaker was implanted, a cardiac electrophysiologic study was

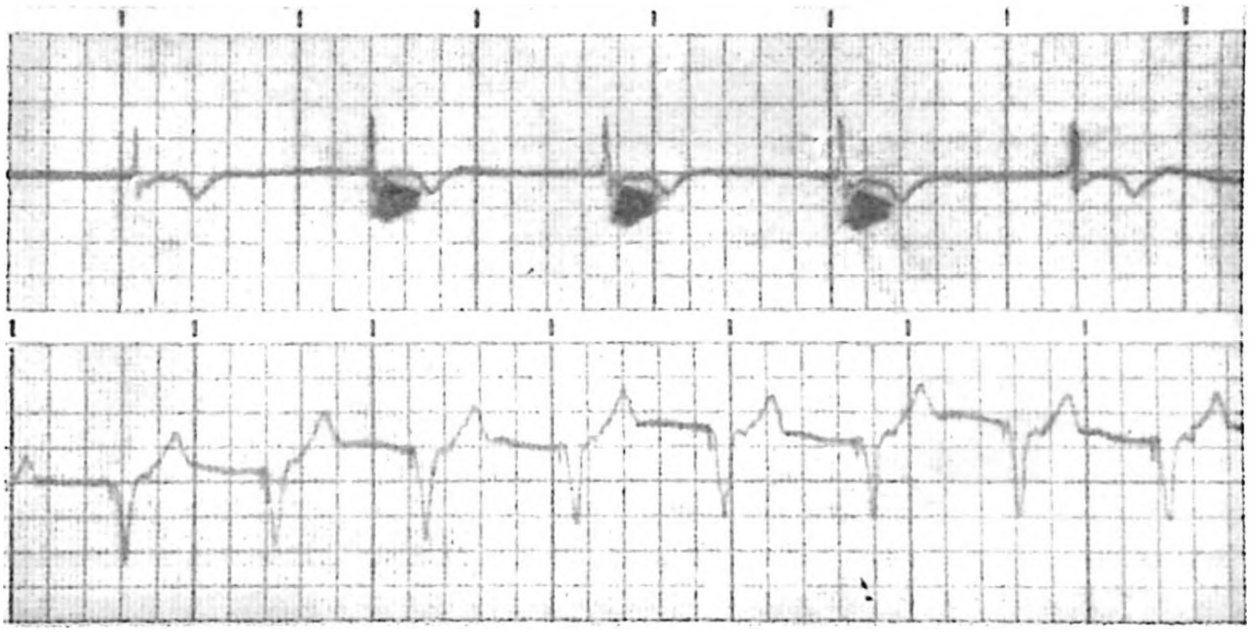

Fig. 2: A low junctional rhythm is noted on the first ECG. Retrograde P waves follow the QRS complex (black arrow). Second panel shows a pacemaker rhythm which was achieved with a VVIR pacemaker.

performed to evaluate sinus node and conduction functions. The study revealed the presence of an abnormal functioning sinus node associated with abnormal A-V conduction time (Fig. 3). A transvenous ventricular rate responsive pacemaker (Telectronics Meta MV) was implanted and adjusted after exercise testing to optimize the child's life style. He showed no signs of dizziness and was discharged on the 20th day of hospitalization.

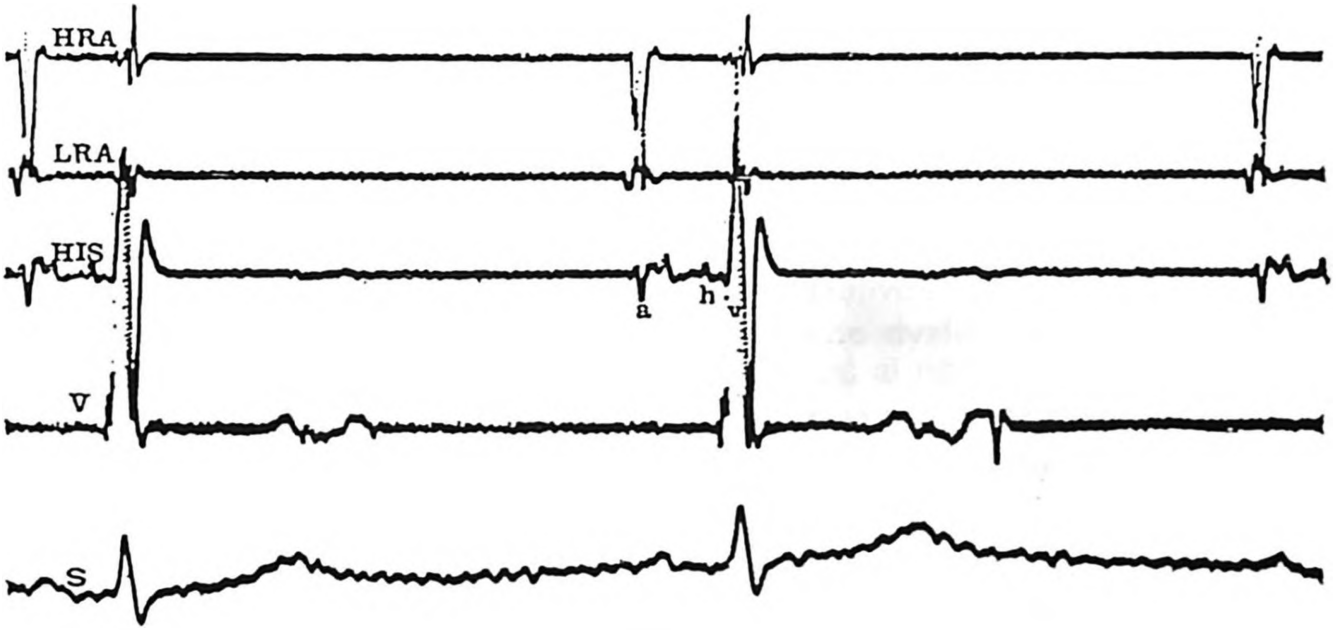

Fig. 3 a:

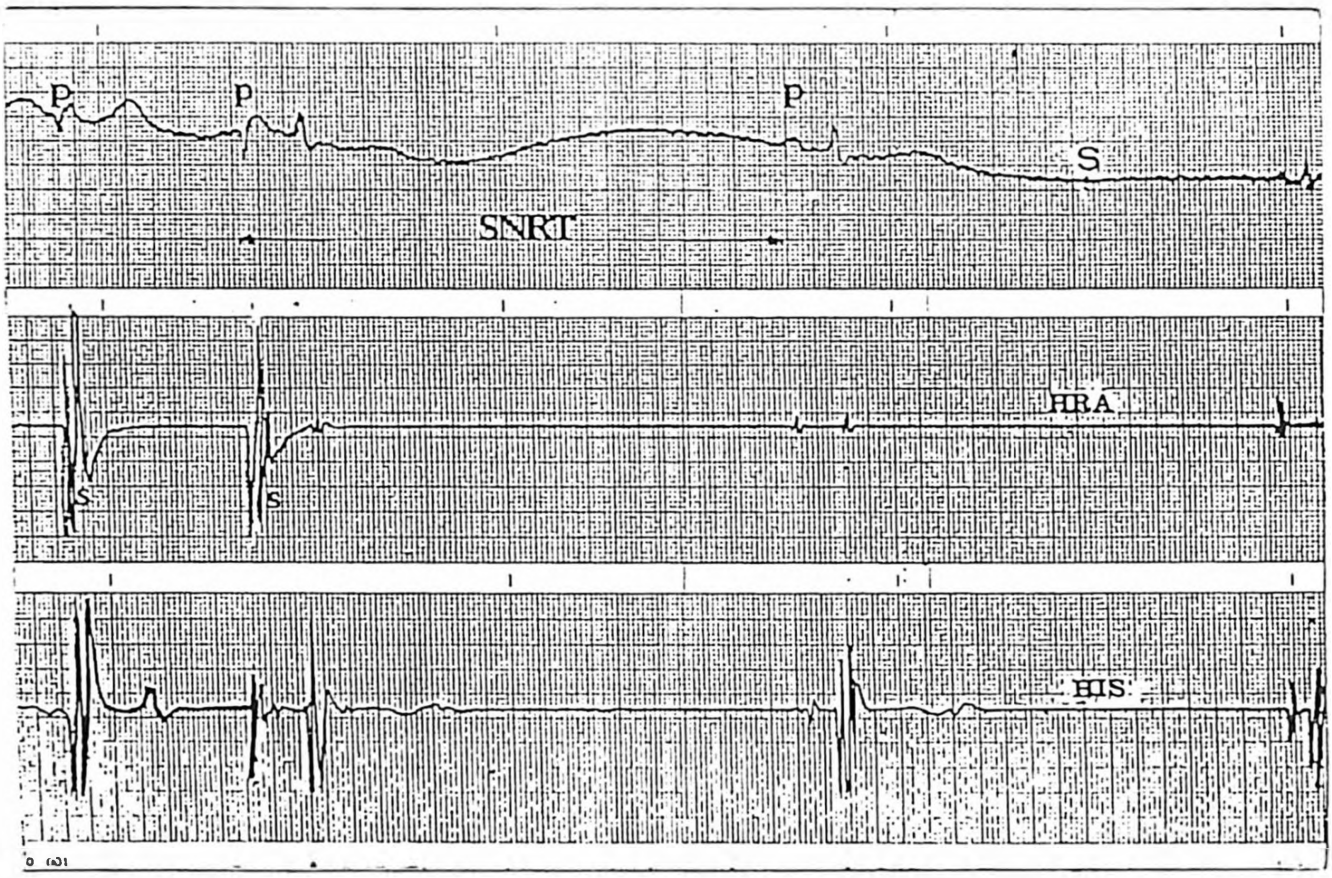

Fig. 3 b:

Fig. 3: Cardiac electrophysiologic study.
a) His bundle recordings revealed abnormally prolonged AH interval (AH = 130 msec).
b) SNRT, which was measured from the surface ECG, was prolonged (SNRT = 2200 msec, CSNRT = 1040 msec).
HRA : high right atrium, LRA : low right atrium, V : right ventricular apex, S : surface.

The other four siblings in the family are healthy. ECGs and exercise tests were normal in these sibs. The parents also evidenced no cardiovascular symptoms.

Discussion

Sinus node dysfunction is an uncommon rhythm disorder in childhood[2, 3]. The disorder occurs sporadically, although it may be frequently noted after cardiac surgery, especially due to the changing atrial anatomy[5].

Since atrial redirection operations have been performed more frequently in recent years, sinus node dysfunction is a relatively rare occurrence in our postoperative care unit. We were able to demonstrate sick sinus syndrome in two siblings, which might indicate a familial occurrence of this disorder.

Dysfunction of the sinoatrial node may manifest itself in several patterns[1, 2]. Etiologies commonly described in this syndrome include cardiomyopathies and surgical injury to the sinoatrial node[5-11]. Some authors have described familial forms of this syndrome[6-11], which may be associated with the fibrosis of nodal or conduction tissue or myocardial disease[6-11]. Allensworth et al[6] found persistent atrial standstill in three siblings and a right atrial biopsy showed interstitial and perivascular deposits of amyloid in one. Although the fibrous degeneration of the sinus node is the main pathologic process in the nonfamilial form of this syndrome, in some patients an associated anomaly of the AV node is also detected[4, 12-13]. These diverse findings may be explained by the varying spectrum of the disease. However there are only a few detailed pathologic studies that discribe the familial form of this syndrome and which suggest that a degenerative process is involved in its etiology[6, 8]. Since we did not perform any pathologic examinations in our patients, we cannot offer any explanation about the etiology of this syndrome. However, we can speculate that fibrous degeneration may be the etiologic mechanism implicated, since pacemaker therapy was successful[9].

It is suggested that the familial sinus node disorder is inherited in a pattern corresponding to an autosomal dominant trait with variable penetrance[6-11]. We are also in accord with this hypothesis because the patients we studied were two siblings.

Since node dysfunction is frequently associated with AV conduction disturbances[1-4]. This concomitat abnormality remains undetected and the surface ECG is within normal limits[1-4]. On the other hand, cardiac electrophysiologic parameters of sinus node dysfunction are not pathologic in certain patients with symptomatic bradycardia which is suggestive of sick sinus syndrome[1-3]. Thus it is recommended that sinus node and AV conduction functions be investigated in these patients so that a precise diagnosis can be established, one allowing for the proper choice of a pacemaker[1-3, 5]. Therefore, we conducted a complete electrophysiologic study in Case No. 2, and we detected abnormal AV conduction in addition to sinus node dysfunction.

In conclusion, given the hereditary character of this condition and the high incidence of conduction disturbances in healthy relatives, it seems advisable to evaluate the family of any patient with sinus node dysfunction, especially of childhood onset.

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